Posterior reversible encephalopathy syndrome mimicking cerebral metastasis: contraindication for biopsy

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Abstract

The posterior reversible encephalopathy syndrome (PRES) is a well described entity of white matter pathology. PRES is triggered by numerous different factors such as acute elevated arterial hypertension, immunosupressive therapy, chemotherapy, etc. The case of a 67-year old woman is presented. The patient was treated for breast cancer 10 months ago and because of acute disorientation a magnetic resonance imaging (MRI) was performed. In the MRI biparieto-occipital hyperintense lesions were seen. Brain metastases were suspected. After chemotherapy and hypertonia and the typical appearance of the lesions in the MRI, PRES was also suspected. Before initializing the surgery for an open biopsy a follow-up MRI had been performed (2 weeks after initial MRI). In follow-up MRI the lesions disappeared completely proving the diagnosis of PRES. PRES can be misdiagnosed as a tumour and surgery could be mistakenly performed. It's important to keep the differential diagnosis of PRES in mind when radiologic features of the syndrome are present.

Introduction

The posterior reversible encephalopathy syndrome (PRES) is a well described entity of a white matter lesion, predominantly involving the posterior portions of the hemispheres.[1] Autoregulatory failure of the blood-brain barrier with endothelial impairment leading to vasogenic edema is the most likely underlying mechanism for PRES.[2] PRES can be triggered by numerous different factors, such as acute elevated arterial hypertension, immunosupressive therapy, chemotherapy, transplantation, eclampsia, renal disease with electrolyte imbalance or systemic infections.[3] Clinical signs at presentation are wide and unspecific. Common symptoms are characterized by alteration of vigilance, seizures, headaches, nausea, and visual disturbances. Radiologic presentation typically indicates bilateral symmetrical white matter edema, in particular affecting the parieto-occipital region and usually sparing the cortical gray matter.[1,4] Early symptomatic conservative treatment is indicated. A follow up magnetic resonance imaging (MRI) after 2-4 weeks is indicated and surgical intervention like attempt to remove the lesion or to perform a biopsy is absolutely contraindicated.

Case Report

We present the case of a 67-year old woman who was admitted in the emergency Department of Neurology due to generalized tonic clonic seizures. After immediate anticonvulsive therapy the seizures ceased. On admission she was hypertonic (190/100 mmHg). She was operated on cancer of the right breast 10 months ago and received adjuvant radio-chemotherapy afterwards. The cranial computed tomography (CT) scan revealed a bihemispheric white matter edema of the parieto-occipital lobe. During her hospital stay she presented decreasing vigilance with delusion. Additionally, as shown in Figure 1A an MRI (FLAIR/T2) was performed, which confirmed biparieto-occipital hyperintense signal alterations in a symmetric pattern. Furthermore, a slightly contrast agent enhancing lesion on the left parietal region was shown. The cerebrospinal fluid (CSF) findings revealed polyomavirus polymerase chain reaction (JC-Virus), which was associated with progressive multifocal leukoencepalopathy (PML). Hereafter the patient was transferred in our neurosurgery unit for biopsy of the lesion in order to determine the exact diagnosis and especially to rule out potential bihemispheric cerebral metastasis with perifocal edema. Pre-operative MRI scan (2 weeks after initial imaging) showed a spontaneous complete disappearance of the previous radiologic pathological findings (Figure 1B). Therefore surgery was cancelled.

Discussion and Conclusions

PRES was first postulated by Hinchey in 1996 and since then there has been a range of more than 1000 publications described its diverse etiology, variety of clinical symptoms and typical neuroradiologic features.[1] The pathophysiology of PRES is controversial. The underlying mechanism of this disorder seems to be a vasogenic autoregulatory dysfunction. Accelerated hypertension and/or cytotoxic agents like chemotherapy apparently induce endothelial damage with extracellular fluid diffusion leading to perivascular edema. However, lack of hypertension does not exclude the possibility of PRES. In 20-30% of cases there is no association with high blood pressure. In the present case, hypertension as well as, chemotherapy as known risk factors were present, although at the time of the disease the patient did not receive chemotherapy.[1,5] Besides the common imaging features of PRES with bilateral white matter edema of the parietal and occipital lobe, lesions can be more extensive, spreading asymmetrically into other structures such as the brain stem, cerebellum, frontal lobes, and sometimes eventually to the basal ganglia or cortex. Moreover, a focal cortical gray matter contrast agent enhancement can be observed as an indication of severe focal impairment of the blood-brain barrier, which has been the case in our patient. In the present case a positive malignoma history of the patient can mislead to a deceptive diagnosis of multifocal cerebral metastasis with perifocal edema. This fact emphasizes the importance of meticulous interpretation of clinical and radiological findings towards PRES as a possible diagnosis, in order to avoid an unnecessary and contraindicated biopsy. The presence of JC-Virus in the CSF also raised PML as differential diagnosis, since neuroradiologic features of this demyelinating disorder can mimick PRES. An early and sufficient symptomatic therapy by lowering blood pressure, anticonvulsive medication and withdrawal of cytotoxic medications is indicated. It is recommended to aim a slow reduction of hypertension in order to avoid possible ischemic events. Antiepileptic drug medication can be usually safely reduced as symptoms and neuroimaging findings resolve. In addition, we recom-

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mend a control cranial CT or MRI 10-14 days after the onset of the clinical disorder. A complete resolution of pathological findings in the MRI with improvement of neurological symptoms within 2 weeks is a strong indicator of PRES. Fortunately, our patient recovered fast without persistent neurological deficits and repeated MRI scan after 2 weeks showed fully regression of the parieto-occipital edema. Although reversible by definition, secondary complications such as ischemic infarction, status epilepticus or even intracranial haemorrhage have been reported. Hence, it is important to recognize PRES as such and begin with proper treatment early to avoid further neurological deterioration or permanent neurological complications.

PRES can be misdiagnosed as a tumour and surgery could be mistakenly planed, especially when it appears after chemotherapy and history of a malignant disease.

It is important to keep the diagnosis of PRES in mind when radiologic features of the syndrome are present.

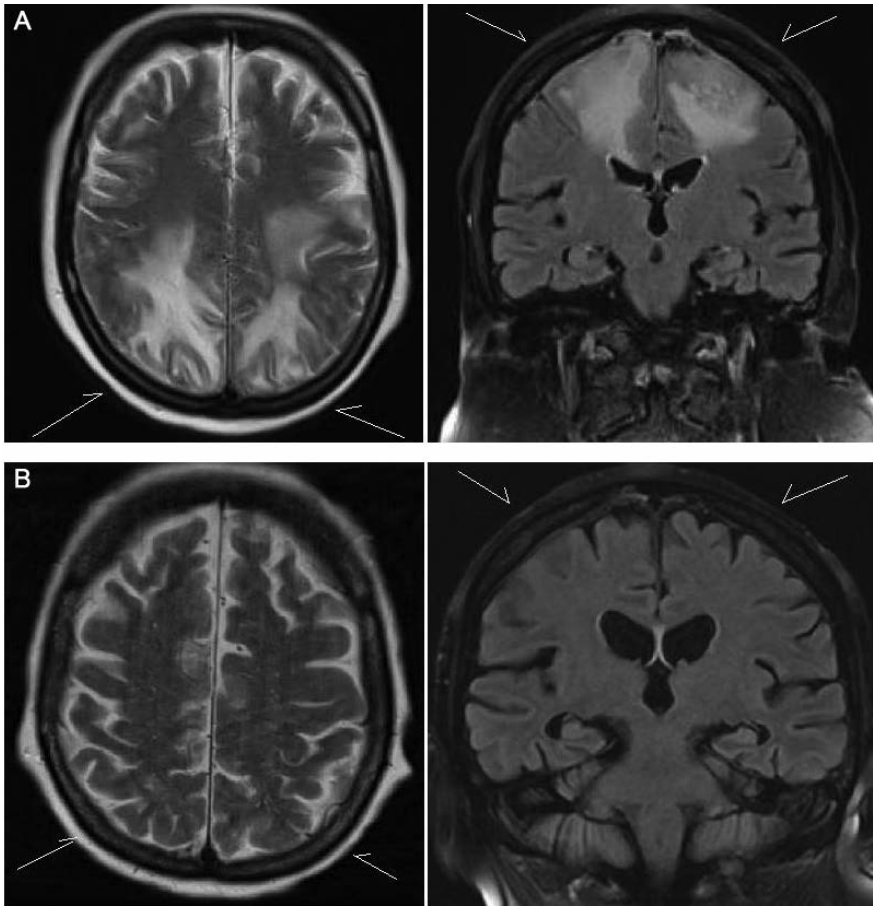

Figure 1. A) Magnetic resonance imaging (MRI) (transversal section, T2) and coronal section, shows the symmetrical biparieto-occipital edema in the white matter (arrows); B) Two weeks after the first MRI a repeated MRI shows complete resolution of the edema (arrows). No tumor can be seen.

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